Pulmonary atresia with intact ventricular septum: medium term follow-up
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Introduction and purpose

Pulmonary atresia with intact ventricular septum (PAIVS) is a rare and complex congenital heart disease characterized by imperforate pulmonary valve and intact ventricular septum. Treatments have been improved over time: surgical techniques have become safer during the neonatal period and percutaneous intervention has been developed as an alternative. This resulted in a reduction in mortality over time, but clinical trend, final cardiac physiology (biventricular 2VR, one-and-a-half 1+½VR, univentricular 1VR), long term complications and therapeutic history can be very complex and currently not well known.

Purpose of this study is to clarify this topic by analyzing a surviving population affected by PAIVS in retrospect.

Methods:
- 59 patients (35 male, 24 female) affected by PAIVS older than 6 years at last evaluation were selected.
- Heart anatomy with echocardiography, angiography and cardiac MRI (when available) was evaluated in the neonatal period (tricuspid valve Detroit z-score, right ventricular morphology, tricuspid valve dysplasia) and during the follow-up.
- Therapeutic history were evaluated (first therapeutic approach, surgical or percutaneous interventions in first year and durring follow-up).
- Final cardiac physiology, clinical outcome (NYHA class, O2 sat) and late complications were evaluated in a median follow-up of 16 years.

Results:

Therapeutic history
- 9 patients (15%) underwent palliation surgery.
- 16 patients (27%) underwent percutaneous right ventricular decompression;
- Palliated patients had lower tricuspid valve (TV) z-score, smaller RV and less TV regurgitation (p<0.05).
- Patients threatened by percutaneous approach had only higher TV z-score (p<0.05) than patients surgically threatened, no difference in RV morphology or TV regurgitation.

Conclusion
- Most of PAIVS patients has reached 2VR physiology at follow-up.
- Patients with final 2VR had higher tricuspid neonatal z-score, more neonatal tricuspid valve regurgitation and more represented right ventricle.
- Therapeutic history is characterized by a lot of interventions.
- After several years a significant amount of 2V patients underwent pulmonary valve replacement for severe pulmonary insufficiency and severe right ventricle dilatation.
- Most patients are asymptomatic at last evaluation.
- Arrhythmias is not an issue.

Statics:

Results are reported as mean±SD or median±SD. T-test for analysis of continous variables and Chi-squared for categorical variables were used.

Results:

<table>
<thead>
<tr>
<th>Nr patients</th>
<th>59</th>
</tr>
</thead>
<tbody>
<tr>
<td>Birth weight (Kg)</td>
<td>3,00±0,58</td>
</tr>
<tr>
<td>Tricuspid z-score</td>
<td>-1,55±1,75</td>
</tr>
<tr>
<td>Tricuspid dysplasia</td>
<td>25%</td>
</tr>
<tr>
<td>Significant TR regurgitation</td>
<td>87%</td>
</tr>
<tr>
<td>RV tripartite</td>
<td>41%</td>
</tr>
<tr>
<td>RV bipartite</td>
<td>44%</td>
</tr>
<tr>
<td>RV monopartite</td>
<td>15%</td>
</tr>
</tbody>
</table>

Last follow-up
- 39 of 50 eligible patients (78%) at last reached 2VR physiology.
- 14 patients have 1 VR, 6 patients 1½ VR physiology.
- Patients with final 2 VR physiology had higher TV z-score, more TV regurgitation and bigger RV at newborn (p<0.05).
- Final 2 VR physiology is not influenced by the type of RV decompression (surgical or percutaneous).
- Most patients are asymptomatic at last follow-up irrespective of final physiology (80% at NYHA I class).
- Only 10% of patients had arrhythmic problems (all supraventricular tachycardia).

Pulmonary issue
- 50% of patients with final 2 VR physiology had severe pulmonary valve (PV) regurgitation during follow-up.
- PV regurgitation is not dependent on first procedure/surgical intervention of RV decompression.
- 12 patients underwent pulmonary valve replacement because of severe RV enlargement.
- RV volume decreased after PV replacement (MRI data average 141±19 ml/mq before, 89±31 ml/mq after).
- Neonatal TV z-score was significant higher in patients who had developed significant RV enlargement (0.37 vs 1.29 , p<0.05).